Chapter 16

Vascular Surgery

Hemangiomas

- Benign tumors of vascular tissue, vascular birthmarks, and vascular malformations are the most common tumors of infancy. They are considered a type of hamartoma and develop from abnormal angiogenesis
- Hemangiomas are usually solitary, located on the head or neck, and occur most often in females
  - Typically appear within a week after birth
  - Superficial lesions are raised, bright red, and bosselated
  - Deep lesions are raised and appear blue-purple
  - The proliferating phase is characterized by 8–12 months of rapid growth
  - Involution occurs over 1–5 years
  - Complete resolution is usually achieved by age 5–7 years
- Classification
  - Neonatal staining ("stork's bite"): light pink lesion on the back of the neck in the midline; usually fades spontaneously
  - Salmon patch: a light pink variety of intradermal hemangioma that sits level with the skin surface, blanches with pressure, and does not change over years. Can be treated with cover-up creams or laser treatment
  - Capillary hemangioma (port-wine stain, nevus flammeus): hyperkeratotic patches (abnormal nerve endings) on the facial skin (deep in the dermis) that are supplied by cranial nerve V (trigeminal). These lesions are permanent (they do not enlarge or regress) and may be associated with Sturge-Weber syndrome (indicating central nervous system involvement). Can be treated with laser ablation
  - Spider angioma: small, central arteriole with a network of radiating intradermal capillaries; these usually appear at age 3–4 years and regress spontaneously

  - Juvenile hemangioma: congenital vascular malformations that usually regress spontaneously after a period of rapid growth. This is a soft, spongy, nontender, reticular pattern of blood vessels in skin over a mass. Treatment consists of observation
  - Strawberry (capillary) hemangioma: these intensely red lesions undergo a period of very rapid growth; complications may develop before regression
  - Congenital arteriovenous (AV) fistulas: multiple and diffuse lesions, 50% of which occur in the head and neck. Treatment consists of intermittent pneumatic compression and complete surgical resection
  - Arterial hemangioma: pulsatile masses that exhibit bruits or thrills and may be associated with sign bleeding or marked regional gigantism. Treatment consists of surgical resection of all AV shunts
  - Venous malformations (cavernous hemangioma): spongy, subcutaneous swellings with a bluish discoloration; these do not regress and may grow to gigantic size, causing disfigurement. Treat with injections of sclerosing agents (eg, tetradecyl sulfate)
  - Kasabach-Merritt syndrome: rapidly enlarging, solitary lesion that presents with hemolytic anemia, thrombocytopenia, and coagulopathy. Treat with interferon
  - Visceral hemangioma: most commonly occurs in the liver and manifests with hepatomegaly, anemia, coagulopathy, and high-output heart failure. Treatment is indicated for large and symptomatic lesions; use steroids, interferon, or embolization
- Diagnose with complete blood count, platelet count, computed tomography scan with intravenous (IV) contrast, magnetic resonance imaging (MRI), or angiogram (rarely indicated)
- Complications
  - Ulceration (most common complication)
  - Bleeding from ulceration
  - Cosmetic concerns
  - Infection
  - Platelet trapping (in Kasabach-Merritt syndrome)
  - Compromise of vital structures (eg, airway, eye)

- Disseminated intravascular coagulation after surgical resection
- Internal organ involvement
  - Liver: hepatomegaly, congestive heart failure
  - Lung: hemoptysis, recurrent pneumonia
  - Treatment for internal organ involvement includes administering prednisone or cyclophosphamide and performing hepatic artery embolization or ligation

- General treatment principles
  - True hemangiomas, if persistent, may require surgical excision
  - Observe uncomplicated hemangiomas
  - Provide compression
  - Sclerosis can provoke an inflammatory reaction, leading to fibrosis and obliteration of vascular channels; inject tetradecyl sulfate
  - Chemotherapy
    - Give steroids (oral or intralesional for periorbital hemangiomas; administer intravenously for life-threatening lesions or when patient's airway or vision are at risk)
    - Isolated cases have shown regression after receiving cyclophosphamide and interferon
  - Embolization can be useful in treating liver lesions and as a preoperative adjunct to surgical resection
  - Surgical treatment is only indicated for complications, including visual impairment, thrombocytopenia, luminal obstruction, uncontrollable ulceration, hemorrhage or infection, atypical growth, congestive heart failure (due to AV fistula), small lesions that can be easily excised, and vascular malformations (do not involute)
  - Laser (argon) treatment is especially useful for port-wine stains, but is not indicated for strawberry capillary or cavernous hemangiomas

Lymphangiomas

- The lymphatic vessels drain protein-rich fluid leaked from capillaries and return it to the blood. Lymph travels through the cisterna chyli, to the thoracic duct, and on to the left internal jugular vein at its junction with the subclavian vein.

Lymphangiomas develop as a result of abnormal embryologic development of the lymphatic system. Unlike hemangiomas, lymphangiomas do not regress spontaneously. Types include cystic hygroma and lymphedema

- Types of benign lymphatic tumors
  - Cystic hygroma (see Chapter 13, Face and Neck)
  - Lymphedema: an abnormal accumulation of lymphatic fluid in interstitial fluid due to abnormal development (aplasia or hypoplasia of lymphatic channels) or obstruction. There are three types: congenital (Milroy's disease; present at birth), praecox (appears in adolescence), and tarda (occurs in middle age)
    - Complications include infection (eg, lymphangitis, cellulitis) and lymphangiosarcoma
    - Diagnose using MRI and nuclear scans (lymphangiograms are rarely performed because of the risk of lymphangitis)
    - Nonsurgical treatment includes diuretics and compression stockings
    - Although staged excision of subcutaneous lesions and skin is sometimes undertaken, surgical treatment is rarely satisfactory

Venous Disorders

- Embryology: right umbilical vein regresses before birth (this may account for a gastroschisis defect occurring to the right of the umbilicus)
- AV malformations
  - Truncal malformations arise from major arterial branches. They are hemodynamically active with large communications, and often form on the upper extremities, head, or neck
  - Diffuse malformations have multiple small communications, are seldom hemodynamically active, and a bruit is common
  - Malformations are bright red, exhibit increased skin temperature, and manifest an audible bruit
  - Complications include bleeding, distal ischemia, and congestive heart failure
  - Diagnose by arteriography
  - Treatment can include surgical excision (high recurrence

rate), compression garments, selective embolization, and proximal ligation (contraindicated, but ligating multiple small feeding vessels may help)

- Congenital anomalies of central veins
  - Duplication of superior vena cava
  - Anomalous pulmonary venous return (total or partial)
    - Infant is cyanotic at birth and has a right-to-left shunt
    - Associated with atrioseptal defect
  - Absence of inferior vena cava, resulting in venous drainage through azygos system; associated with situs inversus
  - Preduodenal portal vein: often associated with duodenal anomalies and malrotation, but also associated with situs inversus
  - Diagnose above disorders with Doppler ultrasound
  - Treat thrombotic complications of congenital anomalies of central veins with heparin anticoagulation or, in the case of acute thrombosis of major veins that is life- or organ-threatening, use thromboplastin

Arterial Disorders

- AV malformations (AVMs)
  - Usually occur in lower limbs and are associated with unilateral limb hypertrophy (hemihypertrophy)
  - A hepatic AVM in a newborn may produce congestive heart failure
  - Intestinal AVM may produce bleeding
  - Physical findings include increased warmth, swelling, and pulsating varicosities
  - Diagnose by Doppler ultrasound, angiography, or MRI
  - Treat by surgical resection (if possible), compression, excision, or angiographic embolization
- Visceral aneurysms (rare)
  - Most common in the renal and splenic arteries
  - Treat by resection and reanastamosis if the aneurysm is >2 cm

Connective Tissue Disorders

- Marfan syndrome
  - Physical findings include tall, thin, body habitus, arachnodactyly and lens dislocation
  - Symptoms include inguinal hernias, spontaneous pneu-

mothorax, pectus carinatum, and aneurysms of the ascending aorta, which results from cystic medial necrosis that ruptures the intima and initiates aortic dissection (sudden aortic insufficiency is a common early manifestation of aneurysm)

- Acquired disorders
  - Kawasaki disease: manifested by skin rash, erythema, edema of the hands and feet, arthritis, cervical adenopathy, and conjunctivitis and aneurysms of the coronary and peripheral arteries
  - Thrombi and emboli: occur in infants of diabetic mothers and manifest with dehydration and polycythemia, which may produce a state of hyperviscosity and result in thrombosis
    - Umbilical artery catheters may be associated with aortic and renal artery thrombosis that leads to hypertension and heart failure
    - Treat with heparin, hydration, and, in some cases, total parenteral nutrition
- Occlusive syndromes
  - Intracranial and extracranial arteries (eg, sickle-cell disease, which is the most common cause of stroke in children)
  - Renal artery stenosis (resulting from fibromuscular dysplasia)
    - Symptoms include hypertension producing headache, visual disturbance, and congestive heart failure
    - Second most common cause of surgically correctable hypertension (coarctation is first)
    - Usually bilateral
    - Diagnose using aortogram with renal angiography
    - Treat with reconstruction using a hypogastric artery graft from aorta to distal renal artery
      - Saphenous vein grafts are contraindicated because of the potential for aneurysmal dilatation when used in children
      - Transaortic balloon dilatation may be effective if stenosis is in branches, but not if stenosis is at a renal orifice (eg, ostium)

Traumatic Vascular Injury

- Due to the laxity of soft tissue, vascular injuries in children may be associated with greater blood loss from third spac-

ing as compared to adults; collateral blood flow is also more extensive in children because of the lack of atherosclerotic narrowing in their vessel lumens

- Vasospasm is common. Carefully perform a thrombectomy with a small Fogarty balloon and continuously flush repair with heparinized (1 unit/mL) saline and papaverine (30–60 mg in 100 cc)
- Computed tomography angiography with 3-dimensional reconstruction may be useful to detect occult vascular injury and avoid conventional angiography
- Catheter-directed angiography is useful for localizing injuries with abnormal physical examinations (ankle-brachial index < 0.9) or soft signs of injury; hard signs of injury (eg, expanding hematoma, pulselessness, bruit or thrill) should be explored
- The greater saphenous vein makes an ideal conduit, although small vessel size may contribute to technical challenges or necessitate a panel or spiral graft
  - Blood vessels will grow with the patient, so an autogenous graft with an interrupted suture line is favored over prosthetic conduits of fixed diameter
  - The most common injured vessels are the brachial, superficial femoral, and popliteal arteries (Tables 16-1 and 16-2)
- A primary end-to-end anastomosis has the best patency and

Table 16-1. Arteries to Ligate Versus Arteries to Reconstruct

Arteries That May Routinely Be Ligated	Arteries That Should Be Reconstructed
Digital, radial or ulnar (not both, preserve ulnar when possible)	Common/internal carotid
External carotid	Subclavian
Brachial (if distal to profunda and adequate signal is present at wrist)	Axillary
Subclavian branches	Brachial (if there is no Doppler signal at the wrist)
Internal iliac and profunda femoral arteries*	Common iliac
	External iliac
	Superficial femoral
	Popliteal
	Tibioperoneal trunk
	Celiac
	Superior mesenteric

*Preserve at least one tibial vessel. The posterior tibial artery is the most critical to repair, followed by the dorsalis pedis and the peroneal arteries, respectively.

Table 16-2. Veins to Ligate Versus Veins to Reconstruct

Veins That May Routinely Be Ligated	Veins That Should Be Reconstructed
Internal and external jugular	Popliteal for prevention of extremity venous hypertension and potential for compartment syndrome
Brachiocephalic	
Left renal	
Infrarenal inferior vena cava (to control damage)	Common and external iliac, if time and patient condition permits
Internal iliac	Portal vein
Tibialis	Right renal vein
Mesenteric	
Subclavian	

can be performed when segmental loss is limited to < 2 cm; a patch repair is an option when > 50% of the native wall is preserved to avoid a residual stenosis

- Classic extremity fractures that contribute to vascular injury are those in the humerus (brachial artery), tibial plateau, and proximal tibia (tibioperoneal trunk)
- A continuous Doppler assessment should always confirm the pulse examination; an ankle-brachial index < 0.9 is abnormal and should be investigated; C-arm and fluoroscopy units should be available for focal vascular visualization, but complete angiography is rarely necessary
- Micropuncture needles with intrinsic guide wires are helpful for emergent arterial and venous access
- Assess grafts for flow every 3 months for 2 years, then biannually for 3 years, and then annually for life
- Intraluminal shunts may be used as a temporizing measure to prevent tissue ischemia, with revision if patient demonstrates symptoms of ischemia
- Revascularization of an extremity after > 6 hours of ischemia necessitates a fasciotomy
- Common technical pitfalls
  - Excessive graft length, resulting in kinking and graft thrombosis
  - Inadequate graft length, resulting in anastomosis disruption
  - Failure to adequately cover the graft with viable soft tissue,

leading to desiccation and disruption
    - A break in sterile technique, resulting in graft infection
    - Inadequate debridement may create an intimal flap, leading to occlusion
    - Poor distal runoff, resulting in occlusion or vasospasm
    - Inadequate immobilization of the associated bony injury, resulting in graft disruption
    - Failure to prevent, recognize, or treat a compartment syndrome after revascularization, resulting in graft occlusion and tissue loss from ischemia
- Postoperative management (all records should include time of observation and name of observer)
    - Frequently monitor distal pulses by palpitation and Doppler ultrasound
    - Assess capillary filling, warmth, and sensation
    - Monitor for hematoma formation
    - Observe for compartment syndrome